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Research Letter

Congenital sick sinus syndrome: Prenatal diagnosis and postnatal follow-up

Jenn-Jhy Tseng [a, *], Ming-Chih Lin [b]

[a] Division of Maternal-Fetal Medicine, Department of Obstetrics and Gynecology, Taichung Veterans General Hospital, Taichung, Taiwan
[b] Division of Pediatric Cardiology, Department of Pediatrics, Taichung Veterans General Hospital, Taichung, Taiwan

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Dear Editor,

Congenital sick sinus syndrome (CSSS) is primarily a disease of the elderly, however, it is also seen in the fetus, infant, and child without heart disease or other contributing factors [1,2].

A 32-year-old woman, gravida 2 para 1, was referred for prenatal care at 36 weeks' gestation because of persistent fetal bradycardia for weeks. A detailed ultrasonographic examination (Voluson 730, General Electric Medical Systems, Kretztechnik, Zipf, Austria) did not find any structural abnormalities in the fetus. Serial fetal echocardiographic studies showed a persistent pattern of slow, regular atrial contraction with 1:1 atrioventricular (AV) conduction (93–107 bpm) (Fig. 1a–c). No blocked atrial extrasystoles or tachyarrhythmia were displayed. CSSS was impressed *in utero*. The fetus was otherwise in a well-being status. Furthermore, all

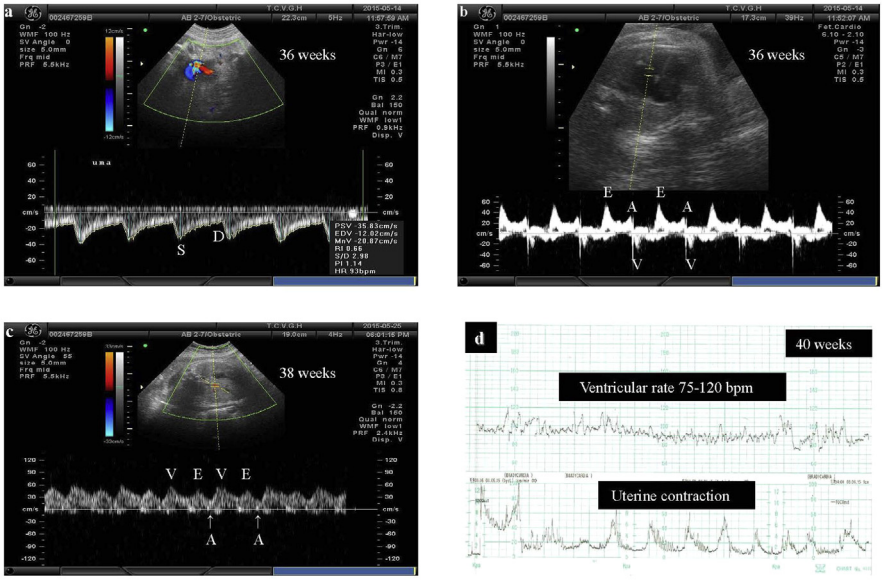

Fig. 1. Prenatal diagnosis of congenital sick sinus syndrome. (a) Pulsed Doppler insonation of the umbilical artery showed a systolic (S)/diastolic (D) index of 2.98 at 36 weeks' gestation. Fetal heart rate (FHR) was 93 bpm. (b,c) Pulsed Doppler insonations sampled at the mitral-aortic continuity and the inlet of ductus venosus at 36 and 38 weeks' gestation, respectively, showed sustained bradycardia with regular rhythm and 1:1 atrioventricular conduction. (d) Intrapartum fetal monitoring showed a decrease in baseline FHR (<110 bpm) with moderate variability. The ventricular rate was 75–120 bpm. A, atrial contraction during late diastole; E, early diastole; V, ventricular systole.

* Corresponding author. Department of Obstetrics and Gynecology, Taichung Veterans General Hospital, 1650 Taiwan Boulevard, Sect. 4, Taichung 40705, Taiwan. Fax: +886 4 23741258.
E-mail address: t1018@vghtc.gov.tw (J.-J. Tseng).

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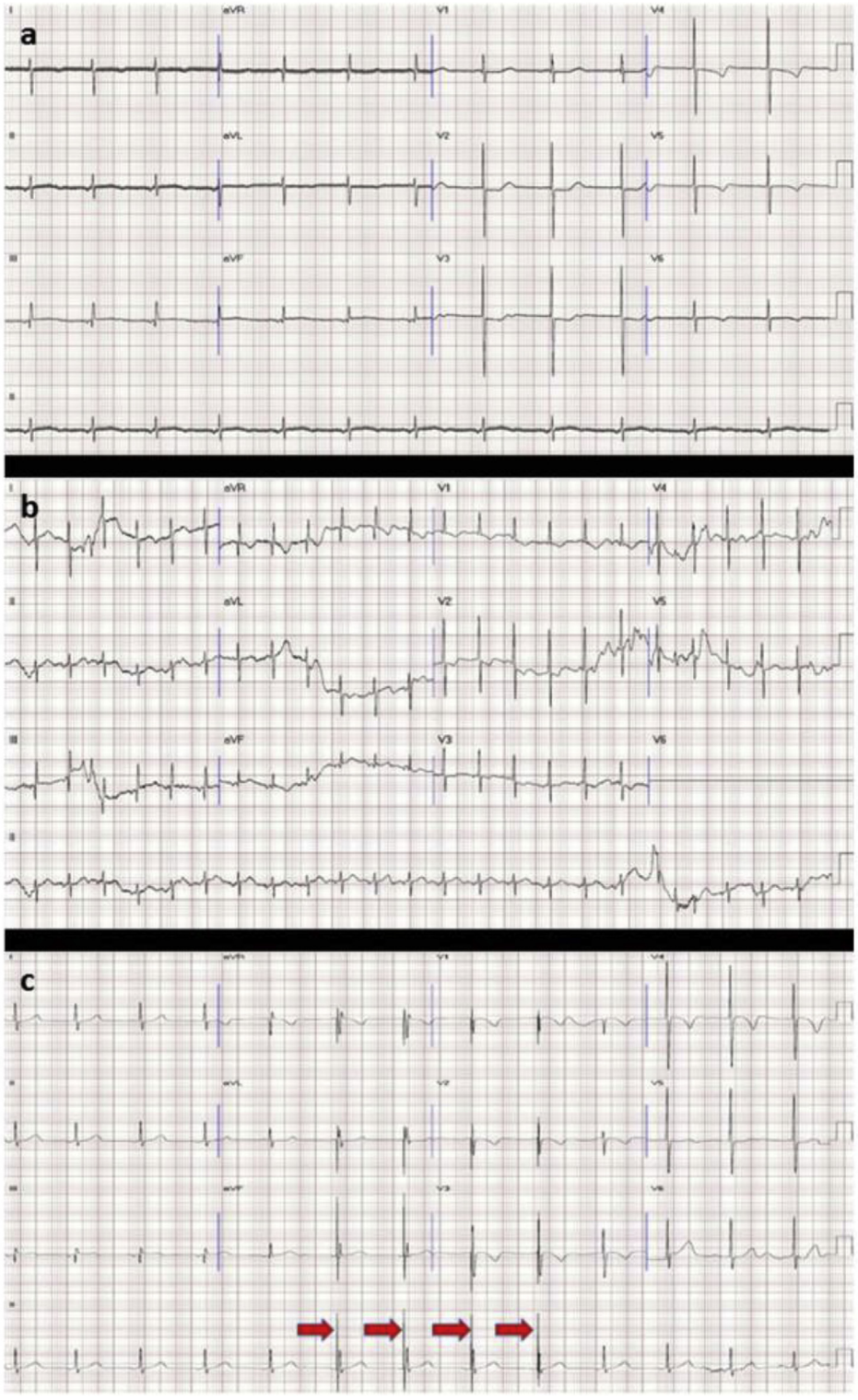

Fig. 2. Postnatal follow-up of congenital sick sinus syndrome. (a) The initial electrocardiography revealed narrow QRS complex without distinguishable p wave. The ventricular rate was 70 bpm. (b) One week after pacemaker insertion, an ectopic atrial pacemaker arose and took over the work of the artificial pacemaker. The p waves were inverted at inferior leads (II, III, and AVF). (c) At 7 months of age, the ectopic natural atrial pacemaker became weaker and the artificial pacemaker worked intermittently.

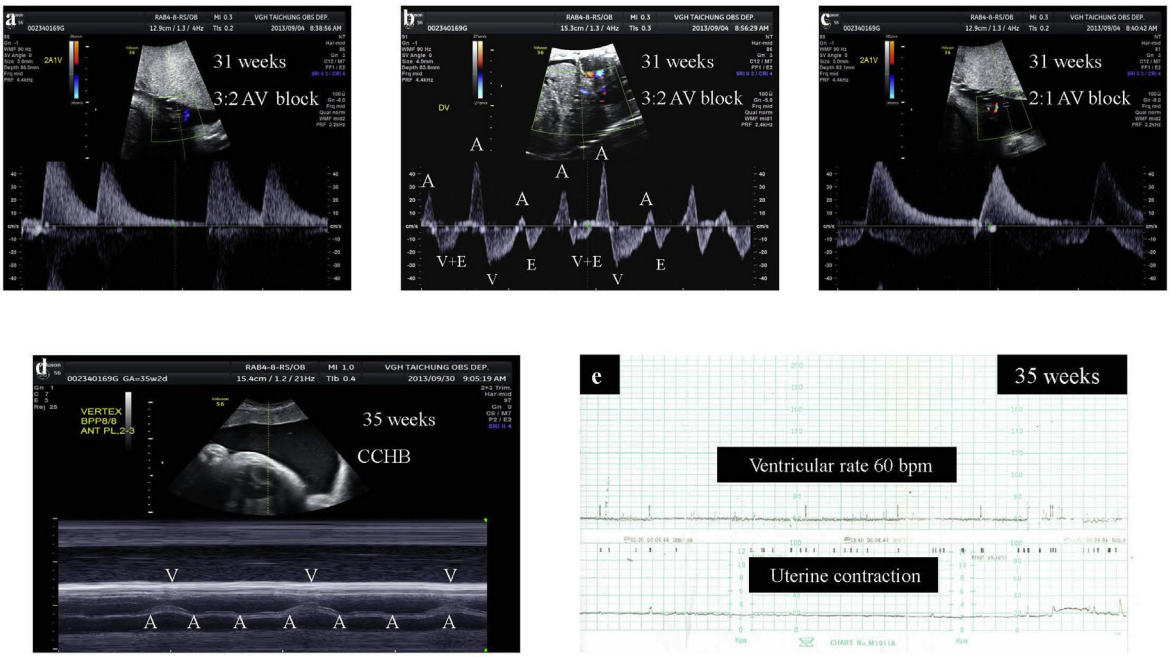

Fig. 3. The progression of second-degree atrioventricular (AV) block to congenital complete heart block (CCHB). At 31 weeks' gestation, Pulsed Doppler waveforms of the (a) umbilical artery and (b) ductus venosus depicted 3:2 AV block. (c) 2:1 AV block was also intermittently found in the umbilical artery during the same study period. (d) At 35 weeks' gestation, M-mode echocardiography demonstrated a lack of AV synchrony. The atrial and ventricular rates were 138 and 60 bpm, respectively. This was consistent with CCHB. (e) Antepartum fetal monitoring of CCHB showed a pattern of bradyarrhythmia with minimal baseline variability. A, atrial contraction during late diastole; E, early diastole; V, ventricular systole.

maternal autoantibodies were negative. At 40 weeks' gestation, a 2920-g male was delivered vaginally with Apgar scores of 7 and 9 at 1 and 5 min, respectively. During labor course, the electronic fetal monitoring demonstrated a category II fetal heart rate (FHR) tracing pattern. It showed a decrease in baseline FHR (<110 bpm) with moderate variability. The ventricular rate was between 75 and 120 bpm (Fig. 1d).

After birth, the initial electrocardiography revealed narrow QRS complex without distinguishable p wave. The ventricular rate was 70 bpm (Fig. 2a). Due to unstable vital signs, a permanent pacemaker (VVIR mode) was inserted via epicardial approach. One week after pacemaker insertion, an ectopic atrial pacemaker arose and took over the work of the artificial pacemaker. The p waves were inverted at inferior leads (II, III, and AVF) (Fig. 2b). However, at 7 months of age, the ectopic natural atrial pacemaker became weaker and the artificial pacemaker work intermittently (Fig. 2c). The diagnosis of CSSS was therefore confirmed. The baby is currently growing and thriving at 12 months of age.

Fetal bradycardia is defined as FHR baseline less than 110 bpm [3]. CSSS, nonconducted atrial premature beats, AV block and congenital long QT syndrome were the major mechanisms for fetal bradycardia [4]. By serial investigations *in utero*, sustained bradycardia associated with regular rhythm and 1:1 AV conduction pattern in our case basically excluded the possibilities of AV block, nonconducted atrial premature beats and long QT syndrome. During labor course, the electronic FHR monitoring showed a decrease in baseline FHR (<110 bpm) with moderate variability. However, the baby was born vaginally and uneventfully.

Congenital complete heart block (CCHB) in the fetus is also expressed as sustained bradycardia and worthwhile to be stated briefly. The FHR in CCHB is lower than CSSS and the pattern of AV conduction is dissociated. Fetuses with CCHB and structurally normal hearts are usually associated with positive maternal autoantibodies (anti-Ro, SS-A or anti-La, SS-B) [5]. Rarely, the progression from second degree AV block could be identified (Fig. 3a–d). Different from that of CSSS, FHR monitoring of CCHB showed a pattern of bradyarrhythmia with minimal baseline variability (Fig. 3e). Postnatal pacemaker implantation is indicated.

The persistence of fetal sinus bradycardia noted during the third trimester may suggest a high possibility of CSSS after birth [4]. An accurate diagnosis of fetal bradycardia would provide the prediction of prognosis and necessity of postnatal management.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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